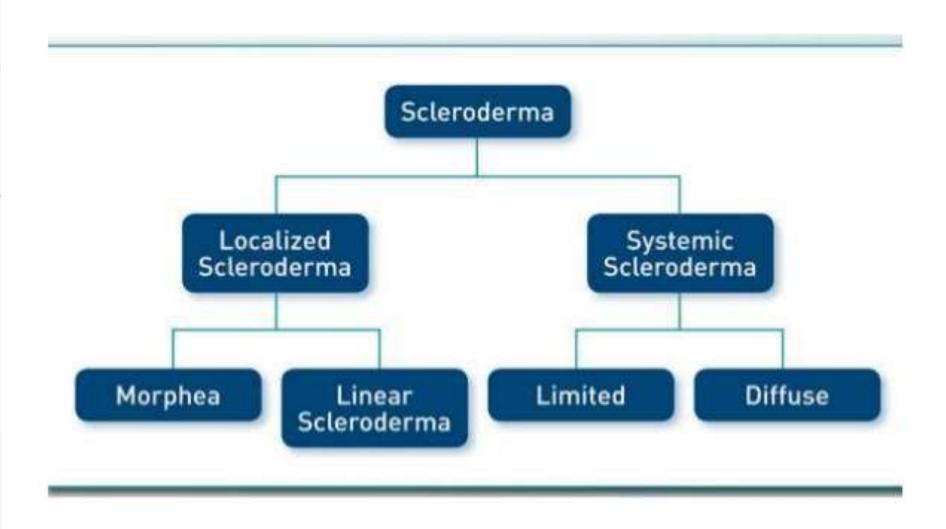
SCLERODERMA

INTRODUCTION

Scleroderma, or systemic sclerosis, is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases. The word "scleroderma" comes from two Greek words: "sclero" meaning hard, and "derma" meaning skin. Hardening of the skin is one of the most visible manifestations of the disease.

CLASSIFICATION



ETIOLOGY

ENVIRONMENTAL FACTORS:

Silica dust

Organic solvents

PVC

Biogenic amines

- **■** GENETIC PREDISPOSITION
- **DEFECTIVE IMMUNOREGULATION**

- Cell mediated immunity CD4/CD8, cytokines
- > Humoral immunity

Hypergammaglobulinemia

Autoantibody production

Antinuclear antibody[+]>95%

• RISK FACTORS:

Genetics. People who have certain gene variations appear to be more likely to develop scleroderma.

Environmental triggers. in some people, scleroderma symptoms may be triggered by exposure to certain viruses, medications or drugs. Repeated exposure — such as at work — to certain harmful substances or chemicals also may increase the risk of scleroderma.

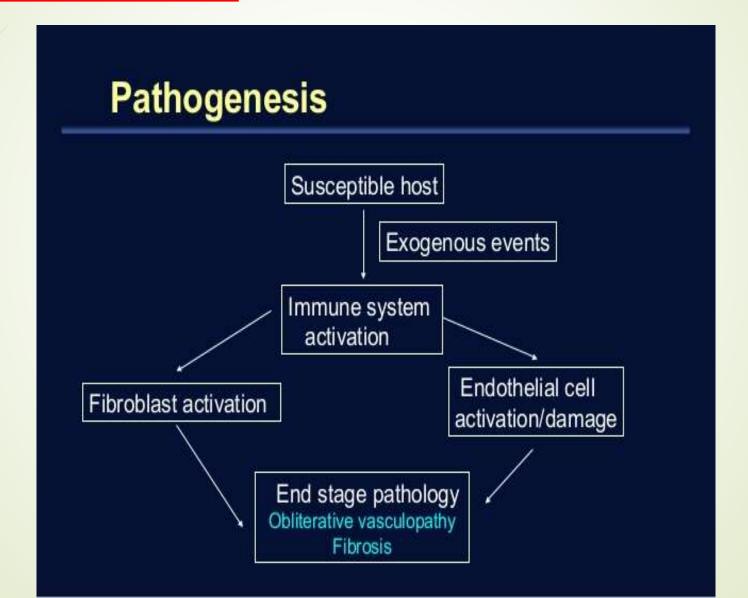
Immune system problems. Scleroderma is believed to be an autoimmune disease. This means that it occurs in part because the body's immune system begins to attack the connective tissues. In 15 to 20 percent of cases, a person who has scleroderma also has symptoms of another autoimmune disease, such as rheumatoid arthritis, lupus or Sjogren's syndrome.

CAUSES:

Scleroderma results from an overproduction and accumulation of collagen in body tissues. Collagen is a fibrous type of protein that makes up your body's connective tissues, including your skin.

Doctors don't know exactly what causes the abnormal collagen production to begin, but the body's immune system appears to play a role. Most likely, scleroderma is caused by a combination of factors, including immune system problems, genetics and environmental triggers.

PATHOGENESIS



COMPLICATIONS:

Fingertips. The variety of Raynaud's disease that occurs with systemic scleroderma can be so severe that the restricted blood flow permanently damages the tissue at the fingertips, causing pits or skin sores.

Lungs. Scarring of lung tissue can result in reduced lung function, which can impact your ability to breathe and tolerance for exercise. You may also develop high blood pressure in the arteries to your lungs.

Kidneys. When scleroderma affects your kidneys, you can develop elevated blood pressure and an increased level of protein in your urine. More-serious effects of kidney complications may include renal crisis, which involves a sudden increase in blood pressure and rapid kidney failure.

Heart. Scarring of heart tissue increases your risk of abnormal heartbeats and congestive heart failure, and can cause inflammation of the sac surrounding your heart.

Teeth. Severe tightening of facial skin can cause your mouth to become smaller and narrower, which may make it hard to brush your teeth or to have them professionally cleaned. People who have scleroderma often don't produce normal amounts of saliva, so the risk of dental decay increases more.

Digestive system. Digestive problems associated with scleroderma can lead to heartburn and difficulty swallowing. It can also cause bouts of cramps, bloating, constipation or diarrhea.

Sexual function. Men who have scleroderma may experience erectile dysfunction. Scleroderma may also affect the sexual function of women by decreasing sexual lubrication and constricting the vaginal opening.

